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Role of the SLP in Management of Huntington's Disease:

A Literature Review

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Role of the SLP in Management of Huntington's Disease:

A Literature Review

by

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Dedication

In loving memory of my brother, Ramon Miranda. You have taught me the world.

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Abstract

Role of the SLP in Management of Huntington's Disease:

A Literature Review

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Huntington's disease (HD) is a devastating, autosomal-dominantly inherited and progressive motor system disease that currently is estimated to affect more than 30,000 people in the United States (National Institute of Neurological Disorders and Stroke [NINDS], 2017). Unfortunately, there is no cure for this disease and prognosis is generally poor, with death occurring at around 15-20 years post-diagnosis (NINDS, 2017). Given HD's distinct effect on motor function, most of the literature has centered around pharmacological treatment for movement. As a result of the disease's progressive nature, cognition, and swallowing are likely to become impaired over the course of the disease; however, the effects of the disease on these systems have been studied to a lesser

extent. Consequently, evidence to support the efficacy of rehabilitative therapies for management of HD symptoms is lacking. Not only is this gap in the literature concerning, as HD carriers could potentially be missing out on therapies that may prove to be beneficial, but it also hinders therapists' use of evidence-based practice for management of the disease. This paper examines available research regarding the management of HD from a communication sciences and disorders (CSD) perspective for use by clinical speech-language pathologists (SLPs). Specifically, this literature review examines evidence-based research regarding the diagnosis, assessment, and management of disorders of speech, language, cognition, and swallowing, with the goal of providing insight into the SLP's role in the management of HD.

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Chapter 1: Huntington's Disease

Huntington's disease is a progressive neurodegenerative and terminal illness that is characterized by distinctive, involuntary, and erratic movements known as chorea (Gudesblatt & Tarsy, 2011). Huntington's chorea was first made well known to the general public in 1872 following a lecture by George Huntington, in which he described the motor disturbances resultant from the disease (Roos, 2010). It was not until the 1980's, when scientists became fully aware of other non-motor symptoms associated with the disease, that its name was changed to Huntington's disease (Roos, 2010). Considered a rare disease, it is estimated that HD affects 5-10 per 100,000 individuals in the Caucasian population and more than 30,000 people in the U.S. alone (Roos, 2010; NINDS, 2017). The average age of onset is around 30 to 50 years of age with an approximated lifespan of 15-20 years post-diagnosis (NINDS, 2017). HD is an autosomal-dominantly inherited disease, meaning that it can be passed down to both male and female children with a 50% chance of inheritance, requiring only one parental carrier of the chromosomal mutation (Roos, 2010). The affected gene, also known as the HD gene, is responsible for production of the Huntingtin protein; however, the purpose of this protein is currently unknown (Gudesblatt & Tarsy, 2011). The mutation occurs on the fourth chromosome and involves excessive reduplication of the cytosine-adenine-guanine (CAG) base of deoxyribonucleic acid (DNA) (Gudesblatt & Tarsy, 2011). The length of CAG repeats inversely correlates with age of onset (Gudesblatt & Tarsy, 2011) and manifestation of HD typically occurs with CAG repeats of 36 or more. This mutation

impedes the proper function of the gene, leading to eventual degeneration of grey matter in the basal ganglia, left striatum, and prefrontal cortex, areas which control motor function, language, and cognition (respectively) (Lambrecq, 2013; Azambuja et al., 2012).

Degeneration in these areas results in a variety of symptoms in HD, including chorea; dystonia, defined as a movement disorder that causes involuntary muscle contracture; dysphagia, defined as a dysfunction of the oral cavity, pharynx, esophagus, or gastroesophageal junction during swallowing; muscle rigidity; difficulty executing fine motor movements; trouble producing speech; cognitive deficits; changes in personality or behavior (depression, suicidal thoughts); and hallucinations (Mayo Foundation for Medical Education and Research, 2017; American-Speech-Language-Hearing Association [ASHA], 2017; Roos, 2010). These symptoms often characterize distinct stages of the disease and change with progression.

More specifically, the course of HD can be divided into three stages: preclinical stage, transition stage, and clinical stage. The preclinical stage can be further divided into the at-risk stage and pre-manifest stage. The at-risk stage begins when at least one parent is affected by HD, leading to the child's uncertainty of carriership (Roos, 2010). The at-risk stage ends and the pre-manifest stage begins once it is determined that the child is a carrier (Roos, 2010). During the transition phase, the carrier may develop strong feelings about changes in behavior, movement, and cognition while uncertainty remains (Roos, 2010). Finally, the clinical stage can be further divided into three sub-stages. Neurological, cognitive, and psychiatric symptoms are often the first symptoms of HD

and are observed in the first clinical stage (Roos, 2010). Chorea is the most prominent symptom in this stage; however, activities of daily living remain relatively independent (Roos, 2010). Motor symptoms become more generalized in the second clinical stage and death by suicide may occur (Roos, 2010). During the third and final clinical stage, chorea becomes severe, leading to complete physical dependence on family and eventual death (Roos, 2010). Additionally, although research has long documented the dysfunction and progressive deterioration of speech, language, and swallow in HD, it is unclear how they evolve in relation to these clinical stages. In summation, HD is characterized by a wide variety of symptoms, including movement, behavioral, cognitive, and linguistic disturbances, which vary in prominence over the course of the disease (Gudesblatt & Tarsy, 2011). Currently, there is no known cure or medication to help slow progression of the disease. As such, SLPs play a role in the management of HD and its symptomatology, with the goal of minimizing complications and maximizing quality of life for those affected (Gudesblatt & Tarsy, 2011).

Chapter 2: Diagnosis

In keeping with ethical guidelines, the SLP should always refer the patient to an appropriate licensed medical professional for proper medical examination and diagnosis when HD is suspected, but not otherwise clinically diagnosed. Generally, a definitive diagnosis of HD can be obtained through genetic testing. Motor, psychiatric, and cognitive evaluations as well as brain imaging (MRI) can also aid in the diagnosis of HD (Roos, 2010). Standardized tests such as the Unified Huntington Disease Rating Scale and Problem Behaviour Scale, as well as other measures, can also be used to scale symptomatology, behavior, and perceived impact of the disease on quality of life (Huntington Study Group [HSG], 2015; Craufurd, Thompson, & Snowden, 2001; Roos, 2010). Such diagnostic tools exemplify the progress made in understanding HD. Nonetheless, along with these advancements, it is also important to consider that receiving an official diagnosis may have ethical, medical, and emotional consequences on the patient and their family (Andersson, Juth, Petersén, Graff, & Edberg, 2013). Ultimately, the patient may choose to not receive an official diagnosis (Andersson et al., 2013).

Chapter 3: Description and Management of Specific Symptomatology in HD

CHOREA

Neurodegeneration of the basal ganglia due to HD results in a disorder of movement known as chorea (Lamrecq et al., 2013). Chorea, characterized by random, involuntary, jerk-like muscle contractions that vary in frequency, amplitude, and intensity, is the most common clinical feature and often one of the first symptoms of HD (Foroud, Gray, Ivashina, & Conneally, 1999; Jankovic, 2009; Roos, 2010). The severity of chorea can range from facial twitching to contraction of the limbs and trunk that results in disturbances in gait and balance (Jankovic & Roos, 2014). Emotional, psychological, and environmental stressors can influence the severity of the chorea (Jankovic & Roos, 2014). Even so, studies have shown that people with HD are often unaware of their choreatic symptoms until they begin to negatively impact their life, such as with job performance (Snowden, Craufurd, Griffiths, & Neary, 1998; Sitek et al., 2011). As a result, treatment of chorea in HD should begin by educating the patient and their family about his or her physical limitations (Jankovic & Roos, 2014). Furthermore, as chorea begins to hinder activities of daily living, the patient may choose to begin pharmacological treatment. Currently, Tetrabenazine is one of only two medications approved by the United States Food and Drug Administration for treatment of chorea (U.S. FDA, 2017). At present, it is unclear whether this drug's benefits outweigh its adverse effects, which can include

sedation, insomnia, and depression (Killoran & Biglan, 2014; Jankovic & Roos, 2014). More recently, the U.S. FDA approved Deutetrabenazine for the treatment of chorea; however, it is unclear how the drug compares to Tetrabenazine in regard to reducing chorea and its adverse effects (Frank et al., 2016; HSG, 2006; Geschwind & Paras, 2016; U.S. FDA, 2017). Nevertheless, the approval of this second pharmacological treatment for chorea is evidence of the progress being made toward better understanding and treatment of chorea in HD.

SPEECH AND LINGUISTIC/PARALINGUISTIC DEFICITS

Hyperkinetic dysarthria, a motor speech disorder usually resulting from damage to the basal ganglia, is common in those with HD and may disturb speech by affecting respiration, phonation, and articulation (McCaffrey, 2013). Speech may also become aprosodic, excessively loud, and harsh (Hartelius, Jonsson, Rickeberg, & Laakso, 2010). Patients with HD may demonstrate lower speech rates, an increase in number of pauses, and difficulty repeating single syllables (Skodda, Schlegel, Hoffmann, & Saft, 2014). Additionally, imprecise vowel articulation and excess intensity variations are also speech deficits observed in people with the disease (Rusz et al., 2014). Furthermore, antidopaminergic medication, often used in the treatment of HD, can negatively impact speech by inducing excessive loudness and pitch variations and highlighting problems with speech timing (Rusz et al., 2014). Overall, dysarthria and the side effects of medication may together negatively affect speech production in people with HD.

Even as research has consistently described the speech alterations in HD, few descriptions exist regarding linguistic disturbances or their determinants in HD (Azambuja et al., 2012). Early in the disease, when neural degeneration is confined mainly to the striatum, deficits in impaired lexical retrieval may be observed in HD (Azambuja et al., 2012). Later in the disease, degeneration of the frontal and temporal regions may further disturb language, but it is unclear what specific deficits may arise as a result (Azambuja et al., 2012). In general, linguistic alterations observed in people with HD may include poorer performance in verbal fluency, narrative writing, and oral and reading comprehension (Azambuja et al., 2012). Individuals with HD may present with difficulty processing metaphors, inferences, and comprehending implicit information (Hartelius et al., 2010). These deficits, however, may reflect difficulties with tasks involving complex material rather than frank aphasia (Azambuja et al., 2012).

In addition to the aforementioned linguistic alterations, research has long documented the presence of paralinguistic deficits, including problems with emotion recognition, in people with HD (Henley et. al., 2012; Paulsen, 2011). In particular, processing of the facial expression for disgust has repeatedly been observed to be impaired in those with HD and can be attributed to the underlying dysfunction of neural systems and structures involved in processing of disgust (Aviezer et al., 2009; Sprengelmeyer, Schroeder, Young, & Epplen, 2006). More recently, research has suggested that this impairment extends to the recognition of all negative facial expressions in people with HD (Johnson et al., 2007). However, despite these deficits, studies have shown that patients with HD may be able to successfully identify facial

expressions when embedded in context, supporting the efficacy of training in this area to improve communication (Avizezer et al., 2009). Furthermore, given that such deficits are often seen early in the progression of the disease, emotion processing deficits have the potential to be used as an early biomarker (Paulson, 2011; Henley et. al., 2012). Overall, it is evident that SLPs must first understand the intricacies and progression of speech and linguistic/paralinguistic deficits throughout the course of HD for proper treatment.

Unfortunately, to the author's knowledge, there are currently no studies that describe the efficacy of speech-language therapy for the management of such deficits or preservation of communicative function in HD. In part, this lack of evidence may result from the difficulty in disambiguating speech and language deficits from cognitive deficits. It may also be attributed to the level of attention placed on what are perceived to be greater health risks, such as chorea, behavior, and dysphagia, which arise from HD. Nevertheless, this current gap in the literature highlights the strong need for studies to examine the efficacy of speech-language therapy for the preservation of language in HD. Future directions for such studies may investigate the efficacy of traditional intervention approaches for speech and language in the preservation of such functions and the resultant impact on quality of life in people with HD. In the absence of such evidence, from a clinical expertise standpoint, linguistic management in HD may heavily rely on encouraging the patient to use compensatory strategies for communication.

Compensatory strategies for the speaker can include speaking more slowly, saying one word at a time, repeating/rephrasing, over-articulating, keeping sentences short, and using gestures (ASHA, 2017). Therapy should also target teaching the primary

communication partner strategies regarding how to make personal adjustments to facilitate effective communication. Some examples of personal adjustments include allowing turn-taking during conversation, slowing their own rate of speech, repeating/ reiterating, keeping questions/statements simple, asking one question at a time, using a yes/no question format, asking for clarification or repeating what you think was said in the form of a question, and eliminating distractions (ASHA, 2017).

With advancement of the disease, augmentative and alternative communication (AAC), defined as communication conveyed by any means except verbal output, may help the patient with HD to express themselves (ASHA, 2017). AAC can be unaided, relying on body language and gestures to facilitate or enhance communication (ASHA, 2017). Aided forms of AAC can range from low-tech (e.g., paper and pencil, communication boards) to hi-tech (e.g., speech generating devices which can be programmed to create messages) (ASHA, 2017). Furthermore, some research has proven such AAC devices effective in facilitating communication in those with HD (e.g., Gelsvartas, Simutis, & Maskeliunas, 2016). Overall, in the absence of evidence-based speech and language interventions for people with HD, the use of compensatory strategies as well as AAC may help enhance and facilitate communication.

Furthermore, in order to fulfill our duties in improving communication in people with HD, SLPs must understand the psychosocial issues surrounding the disease. Some research has indicated that patients with HD may perceive a negative impact on communication when their communication partners speak too fast (Hartelius et al., 2010). Conversely, family members and caregivers may stress a lack of conversational depth,

perceived personality changes in their loved one, and lack of eye contact as factors that negatively impact communication (Hartelius et al., 2010). Given the disagreement between the perceived factors negatively impacting communication, it is evident that both the patient and their communication partners may benefit from engaging in clinician mediated open dialogue to discuss such issues. Moreover, given that both the patients with HD and their communication partners identified a need for increased communication opportunities, the SLP should not only help bridge the aforementioned perceived differences, but also help families identify opportunities to engage in qualitative communication (Hartelius et al., 2010). In summary, SLPs must first understand the psychosocial issues surrounding patients with HD and their families as well as our role in counseling to ensure continuity of care and help improve communication in people with HD.

Behavior and Cognition

Behavioral dysfunction is typically one of the first symptoms in HD. Irritability is often the first sign of psychiatric dysfunction; however, depression, anxiety, aggression, obsessions/compulsions, and hallucinations are also behavioral disturbances observed in HD (Roos, 2010). Additionally, suicidal ideation is common in patients with HD (Craufurd, Thompson, & Snowden, 2001). In fact, suicide is a leading cause of death among people with HD, with higher rates than for any other neurodegenerative disease (Halpin, 2012, Arciniegas & Anderson, 2002; Paulsen, Hoth, Nehl, & Stierman, 2005). If the SLP suspects that the patient is presenting with any psychological problems, he or she

may need to refer the patient to his or her doctor for further referral to an appropriate licensed mental health specialist. Furthermore, when working with a patient with HD, the SLP should consider the effects that both behavioral as well as cognitive disturbances may have on motivation and ability to participate.

Cognitive decline is one of the more common symptoms of HD and often precedes observable motor dysfunction (Stout et al., 2011; Roos, 2010). Cognitive symptoms in HD result primarily from dysfunction of the basal ganglia, with deficits in attention, working memory, and executive function (Azambuja et al., 2012). With progression of the disease, frontal and temporal regions involved in visuospatial processing and episodic memory may be affected (Azambuja et al., 2012). In general, speed of cognitive processing may be slowed even in the early stages of HD and simple mental tasks may become more tiring and effortful (Paulsen, 2011). Although individuals with the disease may have difficulty with explicit learning and memory, they may display even greater difficulty with tasks requiring implicit memory, such as driving a car, playing a musical instrument, and even swallowing (Paulsen, 2011). Patients with HD may also demonstrate impairment in estimation of time that can precede deficits in movement by up to 15 years. Moreover, research has suggested that such deficiencies in behavior and cognition in patients with HD may place the greatest burden on their families and are highly associated with functional decline and likelihood of nursing home placement (Hamilton et al., 2003; Nehl and Paulsen, 2004; Williams et. al., 2010). In the absence of evidence-based treatment approaches for HD-related cognitive decline, treatment should focus on maximizing functionality and quality of life.

Furthermore, in order to help improve the quality of life of patients with HD and their families, SLPs must understand our full role in providing support services. Some studies have indicated that health and personal care and social support services are primary unmet needs among patients with HD (van Walsem, Howe, Iversen, Frich, & Andelic, 2014). These findings accentuate the need for SLPs to take a more active role in facilitating support services and identifying resources for HD families. Support may involve locating and informing HD families regarding HD-specific support groups. Locating resources for home-health support may also be beneficial. In summary, it is critical that SLPs take an active role in providing support and resources to help improve quality of life for those affected by HD.

SWALLOW

Through progression of HD, swallowing may become problematic, posing a choking hazard as symptoms of both dysarthria and dysphagia become prominent (Roos, 2010). Motor weakness and incoordination can affect the function of the pharyngeal muscles, heightening the risk of aspiration. General symptoms of dysphagia may include spillage of food, drooling, difficulty chewing, coughing, and lip/tongue weakness (Dalton, Caples, and Marsh, 2011). Some studies have found that, in people with HD, most dysphagia symptoms occurred in the in the preparatory, oral, and pharyngeal phases of swallow and included postural instability, rapid and impulsive consumption of food, and poor lingual control (Heemskerk and Roos, 2011). For these patients, the oral phase is often characterized by uncoordinated swallow, repetitive swallow, and post-swallow

residue (Heemskerk and Roos, 2011). Coughing, choking, and aspiration are often seen in the pharyngeal phase (Heemskerk and Roos, 2011). As is evidenced by the variety of dysfunction that can result from dysphagia, assessment of the different phases of swallow is critical, as these symptoms can be life-threatening.

Given that dysphagia is a leading cause of aspiration, pneumonia, and asphyxiation, improper management of dysphagia can lead to a respiratory infection and even death (Dalton et al., 2011; Pritchard & Jones, 2014). A modified barium swallow will yield a clear picture of the oral and pharyngeal mechanisms during swallowing (Dalton et al., 2011). Additionally, some researchers have identified the need for a validated dysphagia assessment scale specifically for HD-induced dysphagia in order to provide insight into the clinical features of the swallow observed throughout the course of the disease (Heemskerk and Roos, 2011). The Huntington's Disease Dysphagia Scale, is an 11-point severity scale for dysphagia (Heemskerk et al., 2014). Although currently only validated in Dutch, the scale has been proven to exhibit high reproducibility and construct validity scores (Heemskerk et al., 2014). Overall, current assessment methods effectively provide a clinical description of the swallow impairment, crucial for appropriate treatment.

In general, dysphagia management may consist of compensatory strategies, including modifications in food and liquid consistency. Environmental modifications for dysphagia management may include informing the patient or caregiver about food presentation, special utensils and positioning of the patient (Dalton et al., 2011). In regard to HD-specific dysphagia management, some research has proven that controlling

consistency and portion-size as well as implementing swallow patterns, such as chew-swallow-cough-swallow, may reduce the frequency of penetration and aspiration, for as long as three years for some patients with HD (Kagel and Leopold, 1992). Other research has investigated the use of more specific approaches to address some of the dysphagia symptoms seen in people with HD. For instance, some studies have shown that placing a lemon ice bolus on the patient's tongue before presentation of food of varying consistencies may help stimulate and slow oropharyngeal movements in people with HD (Kagel and Leopold, 1992). Nevertheless, what research has failed to prove is the efficacy of strengthening exercises for the improvement of swallow in HD (Adams, 2009). In brief, a variety of compensatory strategies exist that have been proven effective for the management of HD specific dysphagia. Despite this, with advancement of the swallow dysfunction in people with HD, placement of a gastrostomy tube or feeding tube may be indicated as the individual loses total control of swallow function.

Chapter 4: Conclusion

This paper examined the available research regarding the management of HD from a CSD perspective for use by clinical SLPs. Our current understanding of the disease itself has clearly burgeoned since its first description to the general public in 1872. Specifically, research has identified and characterized phases of the disease to better describe its course and progression. Undoubtedly, this has paved the way for the development of current treatment options for those affected by the disease. The recent approval of a second U.S. FDA-approved drug is evidence of the progress being made toward better understanding and treatment of chorea in HD. Additionally, other non-pharmacological developments have resulted in a deeper understanding of the neural structures implicated in HD and their respective disturbances. Moreover, the above studies have documented and described the variety of disorders involving speech, language, cognition, and swallow affected in HD— areas which are well within the SLP's scope of practice. Nonetheless, despite these advancements, evidence provided by this review highlights the lack of research examining the efficacy of speech-language-cognitive-swallow rehabilitation in the management of HD. In conclusion, there is a strong need for studies that investigate the effects of traditional or novel speech-language-cognitive and swallowing treatment approaches on the preservation of these abilities in HD. Furthermore, SLPs must understand the psychosocial issues surrounding patients with HD and their families as well as our role in counseling and support services to ensure continuity of care and help improve quality of life.

References

Adams, C. M. (2009). Improving swallow function in progressive dysphagia associated with huntington's disease. Retrieved from ProQuest Dissertations Publishing. (UMI 1469152)

American Speech-Language-Hearing Association (2017). Huntington's Disease. Retrieved from http://www.asha.org/public/speech/disorders/HuntingtonsDisease.htm#signs

Andersson, P. L., Juth, N., Petersén, Å., Graff, C., & Edberg, A. (2013). Ethical aspects of undergoing a predictive genetic testing for huntington's disease. *Nursing Ethics, 20*(2), 189-199. doi:10.1177/0969733012452686

Arciniegas, D., & Anderson, C. (2002). Suicide in neurological illness. *Current Treatment Options in Neurology, 4*, 457-468.

Aviezer, H., Bentin, S., Hassin, R. R., Meschino, W. S., Kennedy, J., Grewal, S., . . . Moscovitch, M. (2009). Not on the face alone: Perception of contextualized face expressions in huntington's disease. *Brain, 132*(6), 1633-1644. doi:10.1093/brain/awp067

Azambuja, M. J., Radanovic, M., Haddad, M. S., Adda, C. C., Barbosa, E. R., & Mansur, L. L. (2012). Language impairment in huntington's disease.

Arquivos De Neuro-Psiquiatria, 70(6), 410-415. doi:10.1590/S0004-282X2012000600006

Craufurd, D., Thompson, J. C., & Snowden, J. S. (2001). Behavioral changes in huntington disease. *Neuropsychiatry, Neuropsychology, and Behavioral Neurology, 14*(4), 219

Dalton, C., Marsh, L., & Caples, M. (2011). Management of dysphagia. *Learning Disability Practice, 14*(9), 32.

Foroud, T., Gray, J., Ivashina, J., & Conneally, P. M. (1999). Differences in duration of huntington's disease based on age at onset. *Journal of Neurology, Neurosurgery, and Psychiatry, 66*(1), 52-56. doi:10.1136/jnnp.66.1.52

Frank, S., Testa, C. M., Stamler, D., Kayson, E., Davis, C., Edmondson, M. C., . . . Huntington Study Group. (2016). Effect of deutetrabenazine on chorea among patients with huntington disease: A randomized clinical trial. *Jama, 316*(1), 40-50. doi:10.1001/jama.2016.8655

Gelsvartas, J., Simutis, R., & Maskeliunas, R. (2016). User adaptive text predictor for mentally disabled huntington's patients. Computational Intelligence and Neuroscience : CIN, 2016, 3054258. doi:10.1155/2016/3054258

Geschwind, M. D., & Paras, N. (2016). Deutetrabenazine for treatment of chorea in

huntington disease. *Jama, 316*(1), 33-35. doi:10.1001/jama.2016.8011

Gudesblatt, M., & Tarsy, D. (2011). Huntington's Disease: A Clinical Review. *Neurology Reviews, 19*(5), S1-S8.

Halpin, M. (2012). Accounts of Suicidality in the Huntington Disease Community. *Omega: Journal Of Death & Dying, 65*(4), 317-334.

Hamilton, J. M., Salmon, D. P., Corey-Bloom, J., Gamst, A., Paulsen, J. S., Jerkins, S., . . . Peavy, G. (2003). Behavioural abnormalities contribute to functional decline in huntington's disease. *Journal of Neurology, Neurosurgery, and Psychiatry, 74*(1), 120-122. doi:10.1136/jnnp.74.1.120

Hartelius, L., Jonsson, M., Rickeberg, A., & Laakso, K. (2010). Communication and Huntington's disease: qualitative interviews and focus groups with persons with Huntington's disease, family members, and carers. *International Journal Of Language & Communication Disorders, 45*(3), 381-393. doi:10.3109/13682820903105145

Heemskerk, A., & Roos, R. A. C. (2011). Dysphagia in huntington's disease: A review. *Dysphagia, 26*(1), 62. doi:10.1007/s00455-010-9302-4

Heemskerk, A., Verbist, B. M., Marinus, J., Heijnen, B., Sjögren, E. V., & Roos,

R. A. C. (2014). The huntington's disease dysphagia scale. *Movement Disorders, 29*(10), 1312-1316. doi:10.1002/mds.25922

Henley, S. M. D., Novak, M. J. U., Frost, C., King, J., Tabrizi, S. J., & Warren, J. D. (2012). Emotion recognition in huntington's disease: A systematic review. *Neuroscience and Biobehavioral Reviews, 36*(1), 237-253. doi: 10.1016/j.neubiorev.2011.06.002

Huntington Study Group (2006). Tetrabenazine as antichorea therapy in huntington disease: A randomized controlled trial. *Neurology, 66*(3), 366-372. doi: 10.1212/01.wnl.0000198586.85250.13

Huntington Study Group (2015). Unified Huntington's Disease Rating Scale (UHDRS). Retrieved from http://huntingtonstudygroup.org/tools-resources/uhdrs/

Jankovic, J. (2009). Treatment of hyperkinetic movement disorders. *Lancet Neurology, 8*(9), 844-856. doi:10.1016/S1474-4422(09)70183-8

Jankovic, J., & Roos, R. A. C. (2014). Chorea associated with huntington's disease: To treat or not to treat? *Movement Disorders, 29*(11), 1414-1418. doi:10.1002/mds.25996

Johnson, S. A., Stout, J. C., Solomon, A. C., Langbehn, D. R., Aylward, E. H., Cruce, C.

B., . . . the Predict-HD Investigators of the Huntington Study Group. (2007). Beyond disgust: Impaired recognition of negative emotions prior to diagnosis in huntington's disease. *Brain : A Journal of Neurology, 130*(7), 1732-1744. doi: 10.1093/brain/awm107

Kagel, M. C., & Leopold, N. A. (1992). Dysphagia in huntington's disease: A 16-year retrospective. *Dysphagia, 7*(2), 106-114. doi:10.1007/BF02493441

Killoran, A., & Biglan, K. M. (2014). Current therapeutic options for huntington's disease: Good clinical practice versus evidence-based approaches? *Movement Disorders, 29*(11), 1404-1413. doi:10.1002/mds.26014

Lambrecq, V. V., Langbour, N. N., Guehl, D. D., Bioulac, B. B., Burbaud, P. P., & Rotge, J. Y. (2013). Evolution of brain gray matter loss in Huntington's disease: a meta-analysis. *European Journal Of Neurology, 20*(2), 315-321. doi:10.1111/j.1468-1331.2012.03854.x

Mayo Foundation for Medical Education and Research (2017). Dystonia. Retrieved from http://www.mayoclinic.org/diseases-conditions/dystonia/home/ovc-20163692

National Institute of Neurological Disorders and Stroke (2017). Huntington's Disease Information Page. Retrieved from https://www.ninds.nih.gov/disorders/All-Disorders/huntingtons-Disease-Information-Page

McCaffrey, P. (2013). Chapter 14. Dysarthria: Characteristics, Prognosis, Remediation [Syllabi]. Retrieved from http://www.csuchico.edu/~pmccaffrey//syllabi/SPPA342/342unit14.html

Nehl, C., Paulsen, J. S., & Huntington Study Group (2004). Cognitive and psychiatric aspects of huntington disease contribute to functional capacity. *The Journal of Nervous and Mental Disease, 192*(1), 72-74. doi: 10.1097/01.nmd.0000106004.67587.57

Paulsen, J. (2011). Cognitive impairment in huntington disease: Diagnosis and treatment. *Current Neurology and Neuroscience Reports, 11*(5), 474-483. doi:10.1007/s11910-011-0215-x

Paulsen, J., Hoth, K., Nehl, C., & Stierman, L. (2005). Critical periods of suicide risk in Huntington's disease. *The American Journal of Psychiatry, 162*(4), 725-731.

Roos, R. A. C. (2010). Huntington's disease: A clinical review. *Orphanet Journal of Rare Diseases, 5*(1), 40-40. doi:10.1186/1750-1172-5-40

Rusz, J., Klempíř, J., Tykalová, T., Baborová, E., Čmejla, R., Růžička, E., & Roth, J. (2014). Characteristics and occurrence of speech impairment in huntington's disease: Possible influence of antipsychotic medication.

Journal of Neural Transmission, 121(12), 1529.

Sitek, E. J., Sołtan, W., Wieczorek, D., Schinwelski, M., Robowski, P., Reilmann, R., . . . Sławek, J. (2011). Self-awareness of motor dysfunction in patients with huntington's disease in comparison to parkinson's disease and cervical dystonia. *Journal of the International Neuropsychological Society, 17*(5), 788-795. doi: 10.1017/S1355617711000725

Skodda, S., Schlegel, U., Hoffmann, R., & Saft, C. (2014). Impaired motor speech performance in huntington's disease. *Journal of Neural Transmission, 121*(4), 399. doi:10.1007/s00702-013-1115-9

Snowden, J. S., Craufurd, D., Griffiths, H. L., & Neary, D. (1998). Awareness of involuntary movements in huntington disease. *Archives of Neurology, 55*(6), 801-805. doi:10.1001/archneur.55.6.801

Sprengelmeyer, R., Schroeder, U., Young, A. W., & Epplen, J. T. (2006). Disgust in pre-clinical huntington's disease: A longitudinal study. *Neuropsychologia, 44*(4), 518-533. doi:10.1016/j.neuropsychologia. 2005.07.003

Stout, J. C., Paulsen, J. S., Queller, S., Solomon, A. C., Whitlock, K. B., Campbell, J. C., . . . The PREDICT-HD Investigators and Coordinators of

the Huntington Study Group. (2011). Neurocognitive signs in prodromal huntington disease. *Neuropsychology, 25*(1), 1-14. doi:10.1037/a0020937

United States Food and Drug Administration (2017). U.S. Food & Drug Administration. Retrieved from https://www.fda.gov/default.htm

van Walsem, M. R., Howe, E. I., Iversen, K., Frich, J. C., & Andelic, N. (2015). Unmet needs for healthcare and social support services in patients with huntington's disease: A cross-sectional population-based study. *Orphanet Journal of Rare Diseases, 10*(1), 124. doi:10.1186/s13023-015-0324-8

Williams, J. K., Barnette, J. J., Reed, D., Sousa, V. D., Schutte, D. L., McGonigal-Kenney, M., . . . Paulsen, J. S. (2010). Development of the huntington disease family concerns and strategies survey from focus group data. *Journal of Nursing Measurement, 18*(2), 83-99. doi: 10.1891/1061-3749.18.2.83